Supra-maximal verification of peak oxygen uptake in adolescents with cystic fibrosis

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Aim: Several studies have reported a reduced peak oxygen uptake (VO2peak) during incremental exercise testing in patients with cystic fibrosis (CF). The objective of this investigation was to verify the VO2peak attained in traditional cardiopulmonary exercise testing (CPET) in adolescents with CF by using a supra-maximal exercise test.

Methods: Sixteen adolescents with CF (8 boys, 8 girls; mean age 14.6 ± 1.7 yr; FEV1%predicted [range 45–117%]) volunteered in this study and successively performed CPET and a supra-maximal test [Steep Ramp Test protocol (SRT)], using the same equipment, on the same day.

Results: CPET and the SRT resulted in comparable minute ventilation, breathing reserve and heart rate values. One-way repeated measures ANOVA revealed no significant difference in VO2peak/kg between CPET and SRT (38.9 ± 7.4 and 38.8 ± 8.5 mL·min⁻¹·kg⁻¹, respectively; p = 0.81), and good to excellent associations were found for peak exercise parameters between both tests (r = 0.71–0.98; p < 0.01). Peak work rate was significantly higher (~30%) in the SRT compared with CPET (p < 0.01). Bland–Altman plots show no systematic bias for CPET and SRT measurements of VO2peak/kg. No differences were found between CPET and SRT VO2peak values within and between the maximal and non-maximal effort group (p > 0.04) based on the Rowland criteria.

Conclusion: The VO2peak measured in CPET seems to reflect the true VO2peak in adolescents with CF.

Social benefits and peak work capacity in an adult CF population

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The complex UK benefits system, based on the degree of disability caused by disease, is increasingly being scrutinised and this has adversely affected some CF patients. Physical activity is important in CF, offsetting disease limitations and optimizing quality of life and thereby reducing disability: accurate measurement of this may ensure that patients receive the correct level of benefit. As a marker of this, we measured Peak Work Capacity (%PWC0%) (corrected for lung function) using incremental cycle ergometry in 35 CF adults (17 male) and compared it to the level of benefits received and an activity score calculated from a daily activity questionnaire. Ten (29%) led a sedentary lifestyle and 21 (60%) a low active lifestyle, where the most common form of exercise was walking. Of the 34 who had exercise testing, 3 (9%) had marked and 11 (32%) moderate impairment of capacity. Severity of disease (FEV1%) correlated strongly with PWC0% (r = 0.71, p = 0.03), but correlation of benefits with lifestyle (r = 0.18, p = 0.02) and peak work capacity (r = −0.41, p = 0.02) were less marked (see table). This study shows that PWC0% can be used to aid the evaluation of the effect of disease on everyday life, and hence the disability caused. Such a test may aid patients to obtain the correct level of benefit, thereby avoiding needless and distressing government scrutiny.

Breathlessness during exercise in cystic fibrosis

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Introduction: Breathlessness limits exercise in Cystic Fibrosis (CF). Complex disturbances in respiratory mechanics during exercise make it difficult to determine which alterations contribute most strongly to the sensation of breathlessness. This study aimed to examine interactions between breathlessness, metabolic and ventilatory variables, intrathoracic and intraabdominal pressures and neural respiratory drive (NRD), assessed using diaphragm EMG (EMGdi) during exercise. Breathlessness increased progressively during exercise, whereas PTPdi and minute ventilation plateaued at 70% of the total exercise time, indicating neuromechanical dissociation. The intensity of breathlessness increased with the onset of neuromechanical dissociation, with Borg scores being more strongly correlated with EMGdi (r = 0.988, p = 0.001).

Conclusion: NRD was best correlated to breathlessness following the onset of neuromechanical dissociation.